NIH RELAIS Document Delivery

NIH-10098620

NIH -- W1 AC897

PAMELA GEHRON ROBEY
CSDB/NIDR/NIH Bldng 30 Rm 228
30 CONVENT DRIVE MSC 4320
BETHESDA, MD 20892

ATTN:
PHONE: 301-496-4563
FAX: 301-402-0824
E-MAIL:

SUBMITTED: 2001-12-28 05:02:33
PRINTED: 2002-01-02 08:58:30
REQUEST NO.: NIH-10098620
SENT VIA: LOAN DOC
5385326

NIH Fiche to Paper Journal

TITLE:	ACTA OTO-RHINO-LARYNGOLOGICA BELGICA
PUBLISHER/PLACE:	Acta Medica Belgica Bruxelles
VOLUME/ISSUE/PAGES:	1999;53(2):133-6 133-6
DATE:	1999
AUTHOR OF ARTICLE:	Avril MF; Eloy JP; Panosetti E
TITLE OF ARTICLE:	Fibrous dysplasia of the maxillary: a case report.
ISSN:	0001-6497
OTHER NOS/LETTERS:	Library reports holding title, but not vol or yr 0373057 10427367
SOURCE:	PubMed
CALL NUMBER:	W1 AC897
REQUESTER INFO:	AB424
DELIVERY:	E-mail: probey@DIR.NIDCR.NIH.GOV
REPLY:	Mail:

Acta oto-rhino-laryngologica belg., 1999, 53, 133-136

Fibrous dysplasia of the maxillary : a case report

M. F. Avril, J. P. Eloy and E. Panosetti

Department of Oto-rhino-laryngology, Head and Neck Surgery, Centre Hospitalier de Luxembourg, Luxembourg.

Key words. Fibrous dysplasia ; maxilla

Abstract. *Fibrous dysplasia of the maxillary : a case report.* In a 37 year old woman, complaining of right sided rhinorrhea, frontal headache, sneezing and tearing of the eye, fibrous dysplasia was diagnosed based on a radiological "ground glass lesion" of the right maxillo-orbito-ethmoidal area and a hot spot on total body bone scintigraphy. The lesion was resected using a lateral rhinotomy approach with good functional and esthetic result.

Introduction

Fibrous dysplasia is a benign bone lesion characterized by substitution of the normal bone by a fibro-osseous connective tissue representing the different stages of bone metaplasia.

The history of this disease is well described by BERRYLIN (1) : the first report of fibrous dysplasia was made by WEIL in 1922 ; In 1937, MACCUNE & BUCH introduced the concept of a fibro-osseous dystrophic disease and one year later LICHTENSTEIN mentioned a "fibrous dysplasia" or "Jaffe-Lichtenstein disease".

Fibrous dysplasia represents 2,5% of bone tumors and sex ratio dominates in women with a 3 :1 ratio. Fibrous dysplasia is a slow expanding process which usually stops in the ossifying stage. In severe situations, recurrence of the disease can be seen during pregnancy and adulthood. Teenagers and young adults are most commonly affected.

No specific etiology has been published up to this day. Some factors like trauma, previous surgery, dysgenetic pathology may be influencing factors (2).

In the classification of RAMSEY, three forms of fibrous dysplasia are distinguished.

There is no evolution or changing pattern from one form into another. The monostotic form is found in 65% of the cases. Sex ratio is 1:1 and the lesions are found in patients under age of 30. A polyostotic form is encountered in 30% of the cases affecting females more frequently (sex ratio : 3:1). This form can be seen as a single lesion or as multiple lesions in several bones and is frequently diagnosed before the age of 10. Finally, in disseminated forms (5%), multiple bony lesions are associated with cutaneous and endocrinological signs as in the "Mac Cune — Albright Syndrome" (3) : fibrous dysplasia, large irregularly shaped "café-au-lait" spots and precocious puberty.

Diagnosis is normally made by clinical, radiological examinations and can be confirmed histologically after biopsy or resection.

Standard X-Ray and CT Scan findings show a benign lytic lesion which may be more or less transparent (4). The rather typical "ground glass" aspect may be found (Fig. 2). In MRI examination hyposignal T1 associated with T2 iso- or hypersignal is found (5, 6).

A potential risk of malignant transformation is reported, so these patients need a long term follow up (7).

Presented at the Meeting of the Belgian ENT Society, Brussels, March 7th, 1998.

This report concerns a right orbito-maxillo-ethmoidal fibrous dysplasia.

Case report

A 37 years old Caucasian woman, complained of chronic mucosal rhinorrhea, frontal cephalea, sneezing and tearing of the right eye. Medical history was uneventful.

Clinical examination revealed a hypertrophic right inferior turbinate and clear mucosal rhinorrhea.

Axial and coronal CT Scans demonstrated a "ground glass lesion" of the right maxillo-orbito-ethmoidal area. Ophtalmologic findings were normal.

Total body bone scintigraphy by HDP osteoscan (20 mCi) showed a hyperfixation area on the right maxillary bone.

Based on all these observations, fibrous dysplasia was suspected and surgery was planned.

Resection of the bony lesion combined with dacryocystorhinostomy was carried out by lateral rhinotomy (Fig. 1). Primary closure without reconstruction was done. Histological examination confirmed fibrous dysplasia.

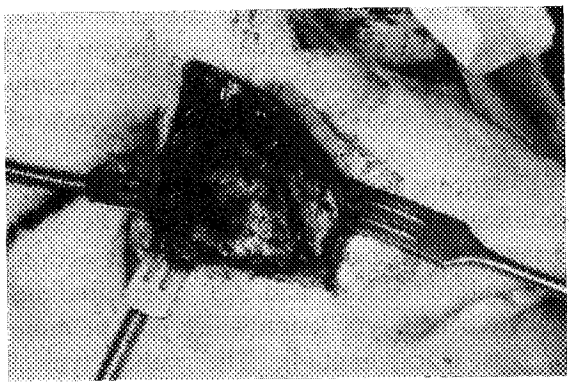


Fig. 1
Surgical approach of fibrous dysplasia by lateral rhinotomy.

After one year, there is no sign of recurrence and the functional and esthetic aspects are fine (Fig. 2).

Discussion

Fibrous dysplasia is a common benign bone lesion. The present illustrates a number of the typical characteristics of the monostotic form of fibrous dysplasia.

Its localisation is in agreement with literature : most frequently in the frontal, sphenoidal, maxillary and ethmoid bones DEROME, only rarely in the temporal and occipital bones.

Clinical signs depend on the localisation and extension of the disease as in the present case, but many cases are asymptomatic. Pain may be related to a hard bulging. In sphenoido-ethmoidal localisations, ophtalmological findings may be encountered (5, 9). Nasal obstruction, tearing, headache, chronical sinus infections may be found in fibrous dysplasia of the paranasal sinuses.

Standard X-Ray and CT Scan findings classically describe a benign lytic lesion which

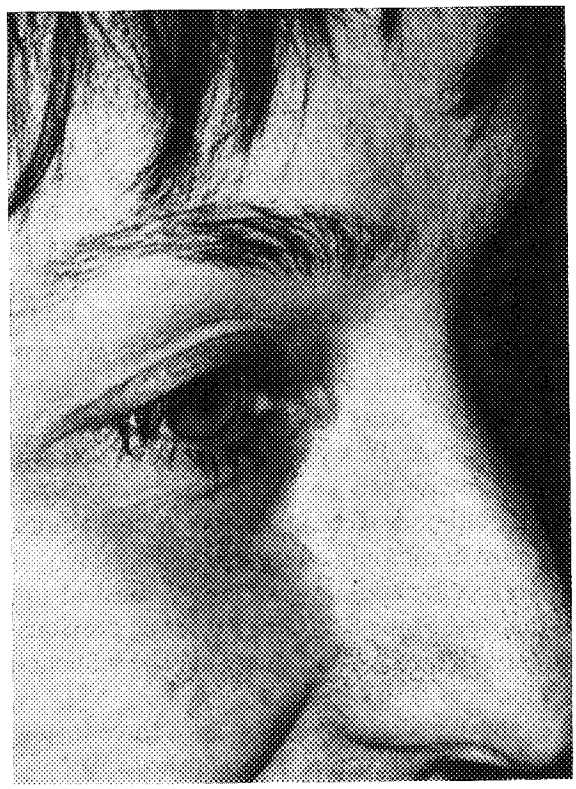


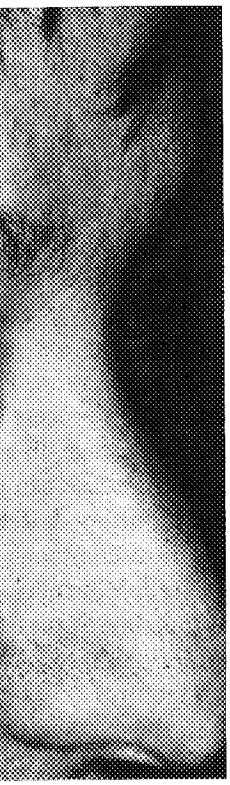
Fig. 2
One year after surgery, the esthetic aspect is fine.

nmon benign bone
es a number of the
ie monostotic form

reement with liter-
the frontal, sphe-
oid bones DEROME,
oral and occipital

on the localisation
se as in the present
symptomatic. Pain
bulging. In sphe-
ns, ophtalmological
tered (5, 9). Nasal
che, chronical sinus
n fibrous dysplasia

CT Scan findings
gn lytic lesion which



esthetic aspect is fine.

may be more or less transparent (4). A "ground glass" aspect may be found (Fig. 2). These characteristics were present in our case.

Bone scintigraphy is important to state the difference between monostotic and polyostotic disease. In the present case there was only one hot spot.

Histology finally proofs the diagnosis of fibrous dysplasia : fibrous stroma and woven bone trabeculae without osteoblasts on the surfaces of the trabeculae (5), as demonstrated for the present case in Figure 3. There was no indication of malignant transformation.

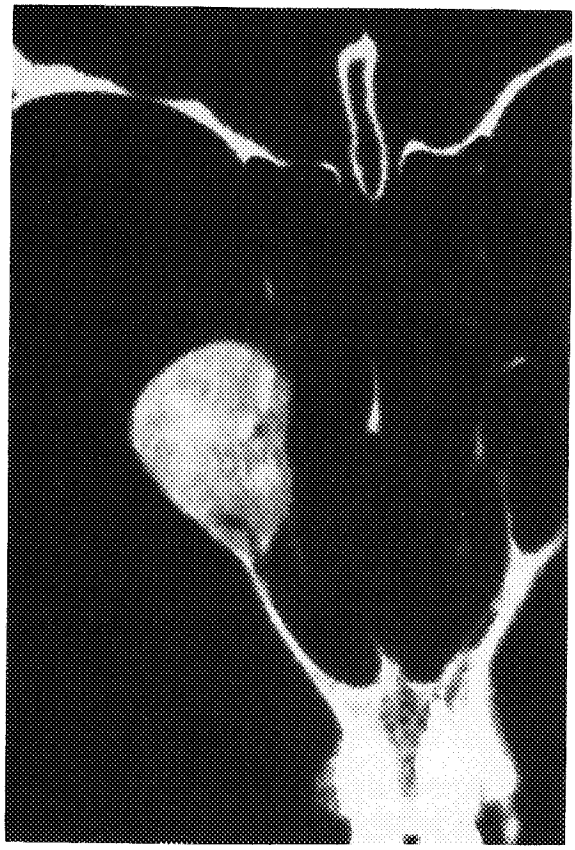


Fig. 3
CT Scan of the parnasal sinuses : specific "ground glass" aspect of fibrous dysplasia in the maxillo-orbito-ethmoidal complex.

The diagnosis of fibrous dysplasia is based on clinical, radiological and histological findings. Histology is the only examination to confirm the final diagnosis. In most cases, this benign lesion is asymptomatic. The choice of

Fig. 4
Histopathology : fibrous dysplasia shows cellulo-fibrous stroma and woven bone trabeculae. A specific sign is the absence of osteoblasts on the trabecular rims (in contrast to ossifying fibroma). HE staining after slow decalcification.

the follow-up depends of functional and esthetic disturbances. If there are no significative symptomatology, clinical and radiological follow-up is necessary every 6 to 12 months. When surgery with or without reconstruction is indicated the most conservative technique should be chosen. Surgical treatment has to be discussed according to the esthetic aspects. In our case report, we performed surgical excision of the lesion via a lateral rhinotomy approach with dissection of the lacrymal system as discribed by Moreau (10). Part of the maxillary bone was removed by osteotomy (11) while drainage of the lacrymal canal was performed in the nasal cavity. Postoperative follow-up was uneventful. One year after the intervention there is a good functional and esthetic result.

Conclusion

Fibrous dysplasia of the maxillo-orbito-ethmoidal can succesfully be treated using a lateral rhinotomy approach combined with a dacryocystorhinostomy.

References

1. BERRYLIN J. F. Fibrous dysplasia of the paranasal sinuses. *American Journal of Otolaryngology*, **15** : 227-230, 1994.

2. Simovic S., Klapan I., Bumber Z., Bura M. Fibrous dysplasia in paranasal cavities : case report. *ORL*, **58** : 55-58, 1996.
3. Schwartz R. A., Spicer M. S. *et al.* Cutaneous fibrous dysplasia : an incomplete form of the McCune Albright Syndrome. *Dermatology*, **192** : 258-261, 1996.
4. Daly B. D., Chow C. C., Cockram C. S. Unusual manifestations of craniofacial fibrous dysplasia : clinical, endocrinological and computed tomographic features. *Postgrad. Med. J.*, **70** : 10-16, 1994.
5. Shapeero G. L., Vanel D., Ackerman V. L. Agressive fibrous dysplasia of the maxillary sinus. *Skeletal radiol.*, **22** : 563-568, 1993.
6. Casselman J. W., De Jonge I., Neyt L., Declercq, D'Hondt G. MRI in craniofacial fibrous dysplasia. *Neuroradiology*, **35**, 234-237, 1993.
7. Deyong X., Hongmei L., Alai Z. *et al.* Spontaneous malignant transformation of fibrous dysplasia. *Chinese Medical Journal*, **109** : 941-945, 1996.
8. Jan M., Dweik A., Destrieux C., Djebarri Y. Fronto-orbito-sphenoidal fibrous dysplasia. *Neurosurgery*, **34** : 544-547, 1994.
9. Moreau S., Bourdon N., Goullet Derugh M. *et al.* Dysplasie fibreuse temporale avec atteinte labyrinthique. *Ann. Otolaryngol. Chir. Cervicofac.*, **114** : 140-143, 1997.
10. Samman N., Piette E., Cheung K. I., Tideman H. Feasibility of osteotomies in fibrous dysplasia of the jaws. *Int. J. Oral Maxillofac. Surg.*, **20** : 353-356, 1991.

M. F. Avril
Oto-Rhino-Laryngology
Head and Neck Surgery
Centre Hospitalier
4 rue Barblé
L-1210 Luxembourg